THE MANAGEMENT OF CHRONIC CONSTIPATION AND RELATED FAECAL INCONTINENCE IN CHILDHOOD

G S Clayden, A S Keshtgar, I Carcani-Rathwell, A Abhyankar


CHILDHOOD CONSTIPATION AND FAecal INCONTINENCE ACCOUNT FOR ABOUT 25% OF PAEDIATRIC Gastroenterologists’ WORK AND ARE WITHIN THE TOP 10 MOST COMMON PROBLEMS FOR GENERAL PAEDIATRICIANS. One way of describing the contribution that members of the therapeutic team make when attempting to help is to follow a typical case history (or patient journey) through the years of evolution and then resolution of the problems. At each stage the likely diagnoses will be discussed together with information on the epidemiology of the more common disorders in the differential diagnosis. The conditions causing difficult defecation in children are similar to a number of childhood problems in the ease with which vicious physical and psychological cycles develop. Unfortunately, this is an area with a shortage of evidence and a plethora of opinion. However, a description of the practice of our team members may provide the reader with valuable insights gained from working with thousands of children with nearly as many variations on the typical constipation theme. Examples of guidelines will be presented with as much supportive evidence as we find. By the end of this article we hope that the reader may feel empowered to set up or contribute to a team that actively manages the problems described with stronger defences against the negative feelings that the chronicity and uncertainties of the problems frequently induce.

NOMENCLATURE
It is impossible to make sense of the literature without being aware of the lack of consensus on the meaning of the words used for the common defecation problems. Attempts have been made to establish a terminology as part of the Rome II consensus on terms in paediatric gastroenterology. However, in practice the terms failed to describe effectively a high proportion of children attending. For the purpose of this paper, the following terms and their definitions are listed in this table 1.

THE EVOLUTION OF CONSTIPATION THROUGH CHILDHOOD
The differential diagnosis of constipation is strongly influenced by the age of the child as illustrated in table 2. This shows the changing patterns of the interaction of structural and psychological factors. The early months with a predominance of structural abnormalities, then the emphasis on dietary problems, with increasing pain avoidance emphasis in the toddler through to the secondary adaptive physical changes and the increasing difficulty with compliance with both medication and lavatory routines in later childhood and adolescence.

DIAGNOSES TO EXCLUDE
Anorectal anomalies are easily missed during a rushed postnatal examination and so careful inspection of the perineum is essential in any baby with constipation. These basic questions need answering:

► Is the anus in the correct position relative to the vulva or scrotum? Degrees of ectopia may be associated with anal stenosis or strictures. A careful per rectum digital examination causes the newborn and infant up to about 6 months no distress if a lubricated fifth finger is used, allowing enough time for pressure on the anus to provoke a recto-anal inhibitory relaxation. Any tightness or stricture is then easily felt as well as an estimate of the volume and hardness of stool in the rectum.

► Could this be Hirschsprung disease? In the rare late presentation of Hirschsprung disease, which should have been discovered as a result of delay in passing meconium, abdominal distension, vomiting or failure to thrive, withdrawal of the examining finger may be associated with an explosive gush of flatus and loose stool. The healthy breast fed baby who may go for longer than a week without passing stool can cause confusion. Normally the breast fed baby passes the usual soft mustard stool, however delayed. Mothers may interpret evening colic as evidence of distress related to these delays and some babies appear to settle after defecation; some, if unwisely subjected to rectal examination, may immediately produce a soft stool. If the breast fed baby is thriving and there is no past history suspicious of intestinal obstruction,
rectal biopsy should be avoided. It is unlikely that rectal biopsy will show the characteristic increase in acetylcholinesterase activity seen in Hirschsprung disease if the onset of even true constipation is later than 1 month of age.4

Anal fissure is relatively common and often associated with very painful defecation. A history of fresh blood seen with the stool or on the nappy or on wiping the bottom is reinforced by the observation of a small skin tag (“sentinel pile”) usually on the anterior limit of the anus (12 o’clock) or posteriorly (6 o’clock). Multiple tags may be seen in association with ectopic “covered” anus and the possibility of anal abuse must always be considered. There is no specific treatment for the acute fissure and usually softening the stool with laxatives and avoiding constipation allows the fissure to heal naturally but relapse is frequent. Rarely excision of a very chronic indurated fissure may allow healing.5 Chronic anal fissure in an older child or a teenager is often associated with chronic inflammatory bowel disease, usually Crohn’s disease, which can be confirmed by histological examination of the fissure or a rectal biopsy.

Prolapse of the rectum is relatively common in young children and may be caused by chronic straining and constipation, protracted diarrhoea, chronic malnutrition as in cystic fibrosis, disorders of sacral nerve innervations as in spina bifida, myelomeningocele, or following repair of high anorectal malformation.7 The underlying pathophysiology leading to rectal prolapse is stretching and weakening of the pelvic floor diaphragm of the anal sphincter muscle complex and suspensory structures of the rectum. The prolapsed rectum may contain lining rectal mucosa, or full thickness muscle wall of rectum, or it may be an intussuscepted sigmoid colon. Recurrent rectal prolapse is another cause of pain related to defecation and may provoke active retention in toddlers old enough to coordinate their anal sphincters and pelvic floor to avoid defecation. Usually episodes of prolapse become more infrequent and of shorter duration with age, with avoidance of constipation and straining on stool for long periods of time.

Occasionally, if nature does not fix the sliding mucosa with intermittent inflammation secondary to the prolapse, surgical treatment is necessary. Injection of sclerosant agents like 3% saline or 5% phenol in almond oil8,9 fixes the sliding rectal mucosa to the underlying muscularis by inducing inflammation and scarring. The injection can be repeated if prolapse recurs. Another frequently employed procedure is a perianal circlage suture described by Thiersch that is inserted around the anus in the submucosal plane and tied over an appropriate size Hegar dilator to tighten the anus.10 Rarely a rectopecty may be indicated which involves removing the coccyx, and re-suspension of the surgically reduced anorectum to the sacrum.11 Laparoscopic repectomy has been reported recently in children with solitary rectal ulcer syndrome and in adults with rectal prolapse, with encouraging results and short term benefits of less morbidity and a reduced hospital stay.12,13

Probably the most dramatic and sudden onset of very painful defecation that would provoke even the most calm of toddlers to avoid defecation is perianal skin streptococcal infection. As this clears rapidly with antibiotics, these should be given early and so inspection of the anus should be mandatory in any constipated child complaining of perianal pain. Less intense perianal inflammation may be seen in cow’s milk intolerance and may suggest an eosinophilic proctitis. Careful exclusion may improve the discomfort but laxatives are usually needed to manage the understandable withholding behaviour. Any form of perianal discomfort is likely to provoke withholding and in those who have a permissively capacious rectum, the delayed stools become hard and large and the fear of painful defecation intensifies.

Rarer diagnoses that should be considered include coeliac disease, Crohn’s disease, hypercalcaemia, hypothyroidism, pelvic or spinal tumours, and chronic intestinal pseudo-obstruction (table 3). However, in these conditions other symptoms and signs would be present. If the child is not

<table>
<thead>
<tr>
<th>Table 2</th>
<th>Differential diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Newborn (1st month)</strong></td>
<td>Infant (1–12 months)</td>
</tr>
<tr>
<td>Anorectal anomaly</td>
<td>Dietary fluid balance</td>
</tr>
<tr>
<td>Anterior ectopic anus</td>
<td>Inadequate intake</td>
</tr>
<tr>
<td>Anal stenosis Hirschsprung</td>
<td>Poor feeding</td>
</tr>
<tr>
<td></td>
<td>Poor feeding</td>
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<td></td>
<td></td>
</tr>
</tbody>
</table>

Table 1  Terms and definitions of defecation problems

<table>
<thead>
<tr>
<th>Term</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Constipation</td>
<td>Difficulty or delay or distress in defecation</td>
</tr>
<tr>
<td>Overflow faecal incontinence</td>
<td>Frequent passage of small volumes of stool in clothing usually without sensation (often referred to as “soiling”)</td>
</tr>
<tr>
<td>Non-retentive faecal incontinence</td>
<td>Episodic passage of normal volumes of stool in socially inappropriate places including clothing (often referred to as “enecrosis”)</td>
</tr>
<tr>
<td>Faecal impaction</td>
<td>Retention of faeces usually palpable per abdomen to a degree that spontaneous evacuation is unlikely</td>
</tr>
<tr>
<td>Megarectum</td>
<td>A rectum of high capacity that empties incompletely or following long delays</td>
</tr>
<tr>
<td>Withholding</td>
<td>An active behaviour resulting in a change in posture that reduces the likelihood of defecation</td>
</tr>
</tbody>
</table>

Hirschsprung disease, usually Crohn’s disease, hypercalcaemia, hypothyroidism, pelvic or spinal tumours, and chronic intestinal pseudo-obstruction (table 3). However, in these conditions other symptoms and signs would be present. If the child is not
thrusting or growing appropriately or there are increasing urinary problems further investigations are essential.

**COMPLICATIONS OF PAINFUL DEFECATION IN THE EARLY YEARS**

Any cause of painful defecation is likely to lead to withholding and contraction of the external anal sphincter and pelvic floor muscles that override the reflex relaxation of the internal anal sphincter in response to rectal filling with stools. Even before infants are in control of these voluntary muscles, a pain provoked withdrawal response causes a virtual spasm. As the child learns to control his voluntary muscles through the toddler stage, more successful withholding of stool is achieved. The parental response to seeing their child in pain and apparently straining to defecate is to conclude that he/she is struggling to pass stool. They may resist the interpretation that the child is withholding, although a careful explanation of the typical straight legged, tiptoe and back arching posture is likely to convince them.

If the child’s retentive efforts are not opposed then there is a risk of impaction of stool in the rectum if the child’s rectal capacity is large enough to allow this. There is no consensus on whether the pathophysiological findings in older children with chronic constipation are merely secondary to persistent rectal loading or whether there is a predisposing megarectum that is present in infancy and permits the original retention.14 As with many other hollow viscera, any form of obstruction will lead to hypertrophy resulting in hyperdynamic smooth muscles of the organ’s walls and an increase in the organ’s capacity. This is not merely of intellectual interest but guides the logic of vigorous early treatment of constipation in the toddler age group to prevent further development of the retentive rectum. The familial tendency to constipation could be explained by familial rectal size, although many other factors such as heightened awareness, bowel related anxiety, and similar diets and exercise patterns must be considered.

**ACTIVE MANAGEMENT**

Early effective treatment will include adequate fibre and fluid intake to encourage the production of soft and bulky stool, but usually a stool softener such as lactulose is needed together with a sufficiently high dose of stimulant laxative such as senna or picosulphate to oppose withholding. No laxative regimen is likely to succeed in the presence of persistent rectal faecal loading. The first phase in all active treatment is to ensure evacuation of the old retained stool. If the child intermittently passes a spectacularly large stool then it is worth waiting until this is passed before starting stimulant laxatives. Prior softening with lactulose, macrogol (Movicol) or docusate may aid this process, depending on the age of the child. Once the rectum is cleared it is vital to start the stimulant laxatives to prevent re-accumulation.

If the stool is not spontaneously passed then more active evacuation methods must be used. This raises another question that is unsupported by clear evidence: is it better to use the oral or rectal route to achieve effective evacuation of impacted faeces. The age and the sensitivity of the child is the main guide here. A toddler who has learned that defecation is painful and should be avoided at all costs because of an anal fissure or perianal skin infection is likely to be terrified of anal treatments and the fears will become intensified. However, an enema performed by an expert nurse with the child effectively sedated may be far less traumatic than trying high doses of stimulant laxative with its resultant abdominal pain or high volume macrogol such Klean-Prep or Movicol that may only be administered by nasogastric tube if drinking the required volume is not possible for a child of that age.

These decisions test our child centred approach where we have a duty to protect the child from abusive treatments at the same time as a need to clear the impacted stool. If the stool is so large that it cannot be flushed from the megarectum, a manual evacuation under a general anaesthetic may be the kindest and most effective method, although risks of anaesthesia may be greater to health than the original constipation. A partially evacuated rectum is likely to reaccumulate stool rapidly and to sabotage the effectiveness of maintenance stimulant and softening laxatives.

The change in liveliness, mood, and appetite that parents report when their child is effectively evacuated conveys them of the benefit of maintaining regular emptying of the rectum. No parents like their child to be on regular nasogastric tube if drinking the required volume is not possible for a child of that age.

**Table 3 Clues to the rarer causes of constipation and soiling**

<table>
<thead>
<tr>
<th>Condition</th>
<th>Clue</th>
<th>Minimal investigation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Coeliac disease</td>
<td>Weight velocity worse than height velocity</td>
<td>Anti-endomysial IgA antibodies as long as IgA concentrations normal</td>
</tr>
<tr>
<td>Crohn’s disease</td>
<td>Abdominal pain and oral ulceration and deep anal fissures</td>
<td>CRP, ESR</td>
</tr>
<tr>
<td>Lichen sclerosus et atrophicus</td>
<td>Usually girl with thickened perianal fissured skin</td>
<td>Consider radioisotope WBC scan</td>
</tr>
<tr>
<td>Hypothyroidism</td>
<td>Height velocity poor</td>
<td>Upper and lower gastrointestinal endoscopy and biopsy</td>
</tr>
<tr>
<td>Pelvic/spinal tumours</td>
<td>Urinary symptoms severe</td>
<td>Dermatological opinion</td>
</tr>
<tr>
<td>Chronic intestinal pseudo-absorption</td>
<td>Upper gastrointestinal symptoms, reflux, failure to thrive</td>
<td>Ultrasound and spinal MRI scan</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Electroagastromy/contrast meal</td>
</tr>
</tbody>
</table>

CRP, C reactive protein; ESR, erythrocyte sedimentation rate; IG, immunoglobulin; MRI, magnetic resonance imaging; TSH, thyroid stimulating hormone.
of waiting rather than actively weaning medication, but it would appear to be unethical to withhold effective treatment to gain this proof.

DEALING WITH CONSTIPATION AND OVERFLOW FAECAL SOILING

With increasing age and social vulnerability, if faecally incontinent, the locus of anxiety gradually moves from the pain of defecation to the unreliability of continence. Placements at nursery may be dependent on faecal continence and certainly by school entry faecal incontinence is a major problem to the child's social integration and self esteem. A careful explanation of the involuntary nature of overflow faecal soiling as being an inevitable consequence of an overfull rectum contracting on its contents with the reflex relaxation of the internal sphincter will help to reduce the blame that is often burdening the child. A key task of the nurse support is to ensure that the child and family understand this physiology and, where permission is granted, to similarly educate teachers and helpers. Although the recto-anal inhibitory reflex is essential for health (this is mediated via the myenteric plexus and its absence is the cause of the obstructive aganglionic segment in Hirschsprung disease), it does lead to the internal sphincter relaxing completely for about 10 seconds during a rectal contraction. The rectum regularly contracts when impacted (contrary to the false belief of rectal inertia in childhood constipation) and so any soft or loose stool in the vicinity of the internal sphincter will leak out, especially as the persistently full rectum produces very little sensation during its contractions. As the leaking stool is of the same temperature as the skin, it is not surprising that the child is unaware of this type of faecal incontinence. This apparent lack of interest in keeping clean is often misinterpreted by parents and teachers as laziness or even delinquency. When challenged, the child is genuinely confused by what he/she could have done to prevent the soiling. With careful explanation, the focus can be redirected to the importance of regular attempts to pass the stool and also to check for soiling and changing underclothes appropriately. Here again nursing advice about the subtle availability of ‘incontinence rescue packs’ at school is essential.

The balance between effective emptying of the rectum and the ability to reach the lavatory in time is particularly difficult if the child needs high dosage laxatives to avoid faecal impaction. The timing of laxatives may help in this age group. Stimulant laxatives given in a single dose each day will hopefully provoke a stool to be passed at a regular time of the day. The timing of the administration of the medication will depend on this pattern and will inevitably vary between individual children. Usually senna is given in the evenings to increase the chance of a stool 24 hours later, whereas picosulfate appears to have a rather more rapid action although there is little concrete evidence for these observations.

The role of psychological factors in the development and maintenance of encopresis is controversial. The evidence available is limited to studies with methodological limitations, thus precluding firm conclusions. While a few studies report poorer social competency and more behavioural problems in children with chronic constipation with or without encopresis than controls, the severity of the psychopathology does not fall in the clinical range. Several studies that have focused on more specific psychological factors in children with encopresis have found that approximately a quarter of children with encopresis reach clinically significant higher scores on antisocial aggressive behaviours before treatment. Hyperactivity and problems with attention, withdrawn behaviour, poor self esteem, and scholastic under performance in reading and spelling have also been reported. This may imply that while approximately 75% of children with encopresis might benefit from a symptom focused behavioural intervention, the remaining children might require a broader intervention. We are currently investigating the psychological characteristics of our referred children with particular emphasis on their social communication skills. While preliminary results indicate increased rates of social communication problems, the results may reflect referral biases to a tertiary service and thus need be replicated in epidemiological samples.

Whether psychological difficulties precede encopresis or whether they are a maintaining factor, it is important to assess and address disruptive behaviours as they are likely to affect the child's compliance with treatment. Refusal to comply with toilet sitting, medications, or clean out procedures undermines the fundamental principles of most treatment protocols. While there are some reports on the effectiveness of psychological intervention in children with encopresis, the evidence is very limited and calls for rigorously designed treatment trials.

DEVELOPMENT OF TREATMENT GUIDELINES

There have been a number of attempts to develop clear and effective guidelines for the management of childhood constipation and soiling from distribution of leaflets through the British Paediatric Association (1994), publication of a consensus document from the USA, Drugs & Therapeutics Bulletin, and the ongoing activity of the working group of the British Society for Paediatric Gastroenterology, Hepatology and Nutrition. There are a number of excellent locally developed packages—for example, “Tough Going” from Scotland. Guidelines will inevitably reflect the available resources locally where the relative scarcity of child and family psychology and psychiatry across the UK limits this essential input. We developed a simple flow chart originally that grew to cover the different approaches with age and to reflect the gradual escalation of intensity with lack of progress. We print the algorithm (fig 1) on one side of an A4 sheet and the advice about alternatives (fig 2) on the reverse.

Everyone developing and evaluating guidelines finds the lack of robust evidence frustrating. The lack of research is not due to complacency or laziness of those working in the field but reflects the intrinsic problems in this particular area such as the lack of clear outcome measures, the strength of opinion that makes allocating a child to a control arm of a comparison with an “accepted” treatment extremely difficult, and the unglamorous nature of the symptoms that makes recruiting researchers and funders more difficult than for life threatening conditions. Our attempt to have a measure of the severity of the symptoms that can demonstrate a change over time is the parent (and/or older child) completed severity score (figs 3 and 4). We devised our own severity score as existing scoring systems were not suitable for this age group and focused on either faecal incontinence or difficulty with defecation. We found that the score correlated with anatomical and physiological features of longstanding constipation and megarectum.
These questionnaires are routinely given to families who complete them while in the waiting room, sometimes with the help of clinic nurses to assist with reading, translation or explanation. Not only does this provide a score but can assist in the consultation when the families have had a chance to view the likely areas of discussion and form a consensus or
Stimulant laxatives
Sodium picosulfate can be exchanged for forms of senna in the flow sheet. May have a more rapid action (12-18hrs) than senna (around 24hrs from ingestion) but this varies with individuals.

Alternative forms of senna:
Senokot syrup (7.5mg per 5ml), senna granules (15mg per 5 ml), senna tablets (7.5mg)
ExLax Senna - each chocolate square contains 15mg

Alternative forms of sodium picosulfate:
Sodium picosulfate elixir. Picolar sachets (heats up when dissolved) also contains Mg
Dulcolax Perles - tablet version of sodium picosulfate

Docuolate
The detergent stool breaking up effect may be matched by the water delivery effect of Movicol and so may be exchanged as a method of softening up a large faecal mass prior to using evacuation methods. Both are likely to increase faecal soiling at this stage.

Alternative forms of docuolate:
Docuolate paediatric syrup (12.5mg/5ml) Docuolate adult syrup (50mg/5ml) Docuolate capsules (100mg)

Bulking agents
These are intended to trap some water to allow colonic contents to be softer and bulkier
Lactulose osmotically increases colonic water and is a probiotic for the normal intestinal flora
Methylcellulose is a convenient tablet form of bulking fibre that can be made into a liquid but this is not available in a licensed form. Soluble fibres such as Benefibra

Macrogols
Movicol can be used to provide more available water in the colon and can be used as twice daily 1-2 sachet of the macrogol reconstituted in 62.5-125ml in the form of Movicol Paediatric Plain sachets (same as Movicol Half but without flavour, ordinary Movicol sachets are reconstituted in 125ml each). The license in childhood is for the use as an intermittent evacuant and can be used according to this table:

<table>
<thead>
<tr>
<th>Day of Evacuation</th>
<th>1st</th>
<th>2nd</th>
<th>3rd</th>
<th>4th</th>
<th>5th</th>
<th>6th</th>
<th>7th</th>
</tr>
</thead>
<tbody>
<tr>
<td>Volume(ml) for 2-4 year olds</td>
<td>125</td>
<td>250</td>
<td>250</td>
<td>375</td>
<td>375</td>
<td>500</td>
<td>500</td>
</tr>
<tr>
<td>Volume(ml) for 5-11 year olds</td>
<td>250</td>
<td>375</td>
<td>500</td>
<td>625</td>
<td>750</td>
<td>750</td>
<td>750</td>
</tr>
</tbody>
</table>

(all volumes can be spread over 12 hours of each day to reduce risk of vomiting)

Alternatives to Movicol
Klean-Prep or Golytely if using high dosage
Irralax that has a different flavour and a slightly different macrogol

Alternative to use of macrogols as evacuant solutions:

Oral route:
Senna or Sodium picosulfate in higher than daily dose if faecal mass only moderately large
Magnesium salts - but likely to produce too much discomfort in children

Rectal route (if tolerated):
 Bisacodyl suppository, Microenema, Phosphate enema

LENGTH OF TREATMENT
As the evacuation phase of the treatment of constipation is the most distressing, prevention of reaccumulation of obstructing faecal masses is a priority. Attempts to artificially wean off laxatives are like to cause relapses. It is advised that dosage should be reduced only when symptoms or side effects of looseness, frequency or urgency of defaecation dictate.

Figure 2 Alternatives to medications or preparations used in the guideline flow chart.
toxin injections with which this was compared in a different trial were similar but to a higher order than in the anal dilatation trial as judged by changes in the severity scores. This has led to a trial comparing intrasphincteric botulinum toxin injections with a similar control group to the anal dilatation trial and an avoidance of myectomy until this trial is completed.
Keeping your information safe

We respect your right to keep information about you private. However, it is very helpful to keep some of this information on computer. This gives me rapid access to results and facts about your problems if you or one of your doctors telephones me for advice. It also allows me to carry useful information about my patients to clinics outside this hospital which often saves families a great deal of travelling.

I keep basic information from these questionnaires on a secure computer (password protected and especially encrypted to prevent unauthorised access). This helps me to watch your progress which helps in decisions about treatments. It is also helpful in research but in this case nothing is published that can in any way identify you. Only members of my secretarial or research team are given some of this information. My research colleagues only collect information on children in the trials for which families have given separate consent.

I send a letter to your referring doctor each time I see you in outpatients. Usually this letter is created from another computer list of information that is also securely protected like the one for the questionnaires. I am happy to show you the information that I keep this way. Only the medical secretaries have access to the information collected from individual clinics. They add your address and your doctors’ addresses; they do not have access to the complete computer file.

I am using the most up-to-date computer and confidentiality protection software that I could find and I am confident that this protects your information effectively.

If you would prefer me not to keep your information this way then I will just read the questionnaire with you in the clinic and I will not keep any of this information on my computer. This will not affect the way we work together apart from any delays in responding to queries arising between visits as we wait for the paper medical records to be found in our stores.

If you have any further questions about this, I am happy to discuss it with you.

Graham Clayden

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**Table 4** Summary of the surgical interventions for patients with severe constipation and/or incontinence

<table>
<thead>
<tr>
<th>Age group</th>
<th>Indication</th>
<th>Surgical differentials</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neonate</td>
<td>Constipation</td>
<td>Hirschprung disease Patient well</td>
<td>Rectal biopsy and wash out, planned pull through</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Patient unwell, other compromising factors</td>
<td>Rectal biopsy and diversion enterostomy followed by staged pull through</td>
</tr>
<tr>
<td></td>
<td></td>
<td>ARM Ano stenosis, perineal fistula, anal membrane</td>
<td>Primary anoplasty/reconstruction</td>
</tr>
<tr>
<td></td>
<td></td>
<td>No perineal opening</td>
<td>Colostomy and staged reconstruction</td>
</tr>
<tr>
<td>Childhood</td>
<td>Constipation</td>
<td>Intra-pelvic mass/lesion</td>
<td>Excision of the lesion as appropriate</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Previous operation for ARM, failure of medical treatment</td>
<td>Anal calibration/dilatation, disimpaction of stool</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Previous surgery for Hirschprung disease, failure of medical treatment</td>
<td>Anal dilatation, botulinum toxin, excision of residual/recurrent aganglionic segment, lateral sphincterotomy/myectomy (and histology), ACE stoma*</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Recurrent anal fissure failure of medical treatment</td>
<td>Excision of chronic anal fissure and histology</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Severe faecal impaction</td>
<td>Disimpaction of stool</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Intra-pelvic mass/lesion</td>
<td>Excision of the lesion as appropriate</td>
</tr>
<tr>
<td>Constipation</td>
<td>Megarectum present</td>
<td></td>
<td>ACE stoma + excision of meagarectum</td>
</tr>
<tr>
<td>and soiling</td>
<td>Faecal incontinence</td>
<td>No meagarectum</td>
<td>Consider ACE stoma</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Previous anorectal malformation ? Megarectum</td>
<td>ACE stoma + excision of meagarectum, anal plug</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Neuropathic bowel + bladder</td>
<td>ACE stoma + Mitrofanoff</td>
</tr>
</tbody>
</table>

ACE, antegrade continence enema; ARM, anorectal malformation.
ROLE OF SURGERY IN THE MANAGEMENT OF SEVERE CONSTIPATION IN CHILDHOOD

Surgery is only used in the most severe cases of chronic constipation and overflow soiling in childhood, but it can play a key role in intractable cases (table 4). With extreme megarectum where there is evidence of sphincter hypertrophy, it is reasonable to consider modifying the strength of the internal anal sphincter. The logic for this approach is as follows:

- where the capacity of the rectum is high, full reflex anal inhibition only occurs at high rectal volumes
- by the time that stools have accumulated to reach this volume, the stools become hard, rounded, and difficult to pass
- without complete emptying of the rectum, the regular rectal contractions are associated with brief (10 second) relaxations of the internal sphincter which leads to passage of loose overflow soiling
- modification of the sphincter should allow more complete inhibition of the internal sphincter at lower rectal volumes and allow more regular complete emptying of the large rectum
- more regular emptying of the rectum encourages a gradual shrinking of the megarectum over time and return of sensory awareness of a full rectum and more effective emptying of stool from rectum.

With this rationale to modify the sphincter being only theoretical, it seems sensible to use a temporary weakening procedure such as intrasphincteric botulinum toxin injections. As with other indications for botulinum toxin injections the muscle weakness lasts between 3–6 months. It should be remembered that even with myectomy, the internal anal sphincter muscle healed and in many cases little evidence of permanent damage is seen in childhood if anal endosonography is checked later.

Where children are dependent on enemas for management of their constipation and faecal incontinence an antegrade continence enema (ACE or Malone) stoma may be very useful. This has been used in children with idiopathic constipation, neuropathic bowel and after surgical repair of anorectal anomaly, Hirschsprung disease surgery, and myelomeningocele. However, we have used it for children with megarectum who have failed to respond to less intensive surgery and full medical and psychological management.

Usually for ACE stoma washouts, the fluid volume is sufficient stimulus to provoke defecation. However, with high capacity megarectum, we add a stimulant to the washout solution in the form of bisacodyl rectal solution diluted in water.

The ACE procedure is not a panacea and is certainly not an alternative or short cut to management that bypasses laxative treatment and full psychological support. A great deal of effort is put in to involve the child fully in the decision to progress to these more intensive treatments. We studied this process of explaining and preparing for the formal consent process in the outpatient clinic setting. It was clear that the triad of the doctor, the child, and the parent was a complex and dynamic relationship when viewing the video recordings of these consultations. We stress to the child that they have a veto over this type of surgical progression that aims to improve the quality of life and how this would differ in life threatening decisions. The age/developmental stage of the child is paramount in their full involvement but if the child does not make clear informed consent, the procedure will be sabotaged and the resulting resentment is likely to aggravate the pre-existing psychological problems.

OUTSTANDING CONTROVERSIES

With the relative lack of evidence base for the management of constipation and faecal incontinence in childhood, there are still many outstanding controversies. The choice of laxatives has been discussed but multicentre controlled trials comparing different combinations of stimulant and softening laxatives at different ages are urgently needed. The importance of food intolerance in the cause and persistence of constipation needs further evaluation. The indications for surgical treatments need further study as well as the best methods for ensuring that children with these non-life threatening conditions have their views heard when invasive treatments are being planned. The interdependent roles of paediatricians, paediatric nurses, child psychologists, child psychiatrists, and paediatric surgeons need more facilitation with organisational arrangements evaluated.

CONCLUSION

Although there have been useful additions to the traditional management of chronic childhood constipation recently such as the early use of macrogol and moving towards using intrasphincteric injections of botulinum toxin rather than vigorous anal dilatation, more evidence is needed for their effectiveness. From our experience in our tertiary referral clinic, it still appears that the greatest shortfall in management across the country is a lack of early supportive medical, nursing, and psychological help. Hope lies in the increasing availability of paediatric nurse led constipation clinics and informed support via the internet such as that supplied by the charity ERIC (now standing for Education and Resources for Improving Childhood Continent) that has widened its remit to embrace faecal as well as urinary continence problems.

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Acknowledgement to Steve Tomlin, Principal Paediatric Pharmacist, Guy’s and St Thomas’ NHS Trust, for his help with the layout of fig 1.

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ARCHIVIST

Bedbugs bite again

A case report from Virginia (Maryanna C Ter Poorten and Neil S Prose. Pediatric Dermatology 2005;22:183–7) illustrates several medical truisms. Firstly, if you don’t remember it you won’t diagnose it. Secondly, non-diagnosis may lead to a farcical situation in which you continue to scratch your head, the patient (in this case) continues to scratch her body, and unnecessary investigations and attempts at treatment may ensue. Thirdly, the patient may, in the end, get the answer from the internet.

A 10 year old girl in Virginia had had a pruritic rash consisting of pink papules and macules on her limbs, buttocks, chest, and neck for 3 months. She had had two punch biopsies and her treatments had included several short courses of oral steroid, antihistamines, antibiotics, and topical permethrin, antibiotic ointments, and steroid creams. The biopsies showed a lot of eosinophils and arthropod bites were thought to be the most likely cause. At this point the child’s mother found some information about bedbugs on the internet. She examined the child’s bed with a torch in the middle of the night and recovered several small brown bugs, which were identified as bedbugs (Cimex lectularius). A pest control service was called in and the child recovered.

Bedbugs were common until the 1940s when they were virtually eradicated by use of dichlorodiphenyltrichloroethane (DDT). Since 1995 increases in bedbug infestation has been reported in the USA, the UK, and Brazil. Bedbugs live in dry corners (mattresses, furniture, floorboards, wallpaper pockets) during the day and emerge at night to feed on human blood. They have never been shown to be vectors for blood-borne disease. Children bitten by bedbugs are most likely to present with papular urticaria.

The recent increase in bedbugs has been attributed to increased air travel and to changes in pesticide management that have eliminated some of the bedbug’s predators such as cockroaches, ants, and spiders. Eradication may be difficult. The services of a professional pest-killing agency may be needed.